A CASE OF HÆMOLYTIC JAUNDICE IN THE NEWBORN*

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ONE feels very diffident, Mr. President, in reporting to this society an unusual case of jaundice in the newborn, remembering the interesting paper presented a year ago by Dr. Hart, on icterus gravis. The case to be reported is, however, different in some respects, and I am led to report it because, judging from the published cases, it is a rare condition. The only other case found was reported in the Dutch literature by E. E. Frank in 1923. His report concerns an infant of forty-four days, and he emphasizes the importance of testing the fragility of the red corpuscles in all cases of prolonged icterus neonatorum as they may prove hæmolytic jaundice.

The case is that of Baby M, male born October 5, 1925, a full term baby, delivered spontaneously, the third child of healthy parents, the father being a young physician. In the family history of neither parent are there any known cases of jaundice, nor have the parents themselves ever been jaundiced. The mother stated, however, that she thought her skin was a little "darker" than usual during the latter part of pregnancy, though not jaundiced. The children of the two previous pregnancies are both healthy boys. The family history is otherwise irrevelant.

The child when born breathed spontaneously, and presented no abnormality to the accoucheur, other than an apparent pallor. The next morning when seen by daylight the child was noted to be intensely jaundiced. The nurse in charge stated, on being questioned, that the child had shown the same jaundice when being carried from the delivery room of the hospital to the nursery, a short time after birth.

Seen at the age of thirty-six hours the baby was quite jaundiced, but appeared otherwise well. It nursed vigorously, cried but little, was afebrile, and took fluids well by mouth. Examination showed the intense jaundice of the skin, conjunctivæ, and the mucous membrane of the mouth, and an otherwise normal physical condition, excepting for a marked enlargement of the spleen, which extended two fingers' breadth below the costal margin. The liver edge was just palpable, the umbilical cord clean and no signs of sepsis were present.

Dr. Graham Ross kindly saw the child with me on the same day, and at his suggestion the blood was examined. The red bloods cells showed 60 per cent normoblasts, 8 per cent megaloblasts and some free nuclei. The white cells showed no striking features. Faint reticulation was observed in a few cells by vital stain. However, increased fragility of the red blood cells to hypotonic saline solutions was found, complete hæmolysis being noted at 0.40 per cent. The bleeding time was not estimated, but was noted to be prolonged.

Examination of the stool gave a positive test for blood, though none was found in the urine, which was a rather brick red in colour, and contained bile salts as shown by a positive Gmelin test. Examination for urobilinogen by Dr. Rabinowitch, at the Montreal General Hospital, showed this pigment to be present in greatly increased amount.

The infant was given 10 c.c. of the father's blood, intramuscularly into each buttock, in an attempt to prevent further bleeding, and subcutaneous injections of saline to increase the urinary excretion, and the breast nursing continued.

There were no further hæmorrhages or unusual events until the seventh day, and no change in the depth of the jaundice. The child continued to nurse well, and was gaining rapidly in weight, the stools were normally pigmented, and the urine still high coloured. On the eighth day of life there was seen slight oozing about the umbilicus at the skin margin, and some small ecchymotic spots on the body and ex-

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tremities, toes, heels, forearms and back particularly. Next day the oozing about the umbilicus had become more marked and was uncontrollable, and a large ecchymosis appeared on the hard palate. Whole blood and hæmostatic serum were given intramuscularly and calcium by mouth and the hæmorrhage from the umbilicus controlled by transfixing the stump with bayonet needles and ligating tightly behind them.

On the ninth day seeping of blood from the various puncture wounds was noted, the child did not nurse as well as formerly, and appeared somewhat exsanguinated, so preparations were made for a transfusion. Professor Waugh, of the Pathological Institute of McGill University, very kindly made a second examination of the blood, of which the chief points of interest are: red blood cells 2,000,000, white blood cells 16,800, hæmoglobin 26 per cent, many nucleated red cells 50 per cent of the red cells showing reticulation and markedly increased fragility. Hæmolysis began at 0.65 per cent and was complete at 0.45 per cent. The normal figures are from 0.45 to 0.30. Examination of the white cells showed many immature myeloid forms. The blood platelets numbered 162,000 per cubic centimeter, and were irregular in size and shape. The bleeding time, though not estimated, was tremendously prolonged.

A transfusion was attempted which was only partially successful, only 50 c.c. of citrated mother's blood being introduced, and the child died shortly afterwards. All the wounds produced in the administration of blood, saline, serum and the umbilical stump were oozing bloody serum before death occurred.

Autopsy was performed fifteen hours post mortem at the Montreal General Hospital, and I am indebted to Professor L. J. Rhea for a report on the findings. The main anatomical features were: intense jaundice of all tissues, enlargement of the spleen, a slight increase of the amount of pericardial fluid, a soft "stone" composed of inspissated bile and amorphous crystals found in the gall-bladder, and the absence of obstruction or abnormality in the biliary system, including the hepatic, cystic and common bile ducts and ampulla of Vater.

Sections were taken from the liver and spleen for microscopic examination.

Regarding the diagnosis of this case we have accepted the definition given by Wilder Tilleston in his monograph on hæmolytic jaundice, *Medicine,* August, 1922, in which he states:

> "For the purposes of this article the term hæmolytic jaundice will be restricted to a form of jaundice, usually chronic, in which diminished resistance of the red cells to hypotonic salt solution is a conspicuous feature, while bile pigment is present in the stools and absent from the urine. Enlargement of the spleen, and anæmia complete the picture. The disease occurs in two forms, the congenital and the acquired, the former being by far the commoner. The congenital type belongs to the inheritable diseases occurring often in several successive generations, occasionally in several members of a family without cases among the ascendants and also in a strictly congenital form, a single member of a family being affected from birth."

Tilleston also mentions that reticulated red cells are present in large numbers.

This case then falls within the definition above in that it presents jaundice, an enlarged spleen whose microscopic picture is that associated with hæmolytic jaundice, increased fragility of the red cells to hypotonic salt solutions and reticulated red blood cells. In cases of jaundice due to obstruction in the biliary system resistance of the red blood cells to hypotonic salt solution is usually increased. In addition the urobilinogen of the urine was increased, which is not suggestive of an obstructive jaundice, but rather of a hæmolytic type. You will recall that the hæmoglobin liberated in hæmolysis is converted by the Kupfer cells of the liver into bilirubin and as such is excreted through the gall-bladder and ducts into the intestinal tract. In the intestine it is converted into urobilinogen, and as such re-absorbed into the circulation and carried back to the liver to be re-excreted as bilirubin. If, however, the amount returned to the liver be in excess, that excess is excreted through the urine. The point to be stressed is that the presence of urobilinogen in the urine excludes the presence of any obstruction in the biliary system. However, in the last number of the *Journal of Experimental Medicine* there appeared an article by McMaster and Elman, April, 1926, showing that urobilinogen may be formed in the liver in the presence of certain organisms, normal inhabitants of the gastro-intestinal tract. This fact, while of interest, probably bears no relation to this case, in that the child was jaundiced from birth. The explanations possible for the increased amounts of urobilinogen in the urine, as in the present case, are two: (a) defective function of the Kupfer cells due to an hepatitis, which is not supported

by the cellular picture of the liver, or (b) an increased liberation of hæmoglobin by hæmolysis.

Regarding the etiology of the disease we know nothing. Tilleston divides hæmolytic jaundice into two types, cryptogenetic and secondary. Our case obviously falls within the first group. Why the child should have bled so freely and uncontrollably we are at a loss to explain. We do know that in some severe infective types of jaundice hæmorrhages do occur. In this case there were no demonstrable signs of infection. It is possible that the jaundice may have been complicated by the presence of hæmorrhagic disease of the newborn.

What type of treatment we should suggest if it were our fortune to see such another case is difficult to state with any assurance of success. Immediate transfusion, or exsanguination transfusion, is certainly indicated, and possibly an early splenectomy.

ABDOMINAL PAIN IN CHILDREN IN EXTRA-ABDOMINAL CONDITIONS*

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PAIN, being a purely subjective symptom, we are dependent for our knowledge of its nature upon the ability of the patient to give us exact information. It follows, therefore, that precise knowledge concerning the nature and severity of pain in young infants must be very meagre indeed, and that unless our methods of examination can be considerably perfected, we shall remain in the dark concerning pain, referred and otherwise, in infants under two years of age, more than to infer from the actions of a child that he is in pain. Even in older children, where attempts are made to localize pain, it is often with the greatest difficulty that exact localization can be made possible. It is easier to elicit tenderness, than to elicit a statement concerning the exact localization of pain. In dealing with pain in its relation to disease in children, therefore, the difficulties which arise seem well-nigh insurmountable. Particularly are the difficulties great when we endeavour to localize a pain to a certain point. The child with a sore throat will, when he complains of pain at all, often say that his neck is sore, or a child with earache, merely that he has pain in the head.

Hilton said many years ago, that when a patient is suffering from pain in any part, he instinctively believes that he must be suffering from inflammation of that part, and he emphasized the fact that pain is not by itself an indication of an inflammatory state, for it may exist without any inflammation of a part complained of. Nothing is more natural than for the patient, or the parents, to assume that the seat of the pain is the seat of the disease, and nothing is more common than to find symptoms of severe pain quite remote from the seat of the disease. Pain does not always depend on disease of the painful part. It depends rather on stimuli reaching the brain from afferent nerve fibres. A child may have pain in his legs, and disease in the spinal cord, or pain in his abdomen, and disease in his chest. One experiences, in practice, so many instances of pain remote from the seat of disease that it has seemed fit to take stock of a few experiences in this connection, especially with reference to abdominal pain associated with disease outside the abdomen. The importance of fuller knowledge of such conditions can hardly be over-estimated, particularly with reference to the abdomen, where one is apt to be misled into believing that acute abdominal disease actually exists, or conversely to overlook a true intra-abdominal con-

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